Case reports

Aortic atresia occurring with complete transposition of great arteries

K M McGARRY, J F N TAYLOR, F J MACARTNEY

From the Thoracic Unit, The Hospital for Sick Children, Great Ormond Street, London

SUMMARY Aortic atresia occurring with complete transposition of the great arteries (ventriculoarterial discordance) has not previously been reported. A patient with this condition is described, who is alive and relatively well at the age of 6 years. This survival contrasts conspicuously with that of patients with the far commoner situation of aortic atresia with normally connected great arteries. It is suggested that this difference in longevity is a result of the greater ability of the left ventricle to supply both the systemic and pulmonary circulations.

Case report

A female Caucasian infant, weighing 3.5 kg, was born by normal delivery after an uneventful pregnancy to non-consanguineous parents on 9 May 1973. Her neonatal progress was unremarkable and no abnormality was noted until the age of 14 months, when a murmur and mild cyanosis were discovered at routine examination. The child was referred for cardiac catheterisation, underwent two inconclusive investigations, and was started on digoxin and frusemide for mild cardiac failure. At the age of 4 years, she was seen at The Hospital for Sick Children. She was complaining of mild exertional dyspnoea and sweating, but otherwise led a relatively normal life, attending normal school. Developmental milestones were normal. On examination, she was below the third centile for height and weight. She was mildly cyanosed and not dyspnoeic at rest. Her heart rate was 140/min and regular. There was no clubbing. The peripheral pulses were of normal volume and synchronous. The jugular venous pulse was elevated with a prominent "a" wave. The anteroposterior diameter of the chest was much increased. The apex beat was to the right and a systolic thrill was palpable at the right sternal border. An ejection systolic murmur and a mid-diastolic flow murmur were audible maximally in the 3rd right intercostal space. The 2nd heart sound was single. There was 4 cm hepatomegaly and the lungs were clear.

INVESTIGATIONS

The chest x-ray film and bronchial tomography disclosed situs solitus of the atria and viscera, a grossly enlarged heart shadow mainly on the right, and increased pulmonary vascularity.

The electrocardiogram showed a normal P wave axis, with right atrial hypertrophy, left ventricular hypertrophy, and interventricular conduction delay.

Table Haemodynamic data

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure (mmHg)</th>
<th>Mean pressure (mmHg)</th>
<th>Oxygen saturation (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>a 28</td>
<td>17</td>
<td>70</td>
</tr>
<tr>
<td>High right atrium</td>
<td>v 20</td>
<td>69</td>
<td></td>
</tr>
<tr>
<td>Mid right atrium</td>
<td>v 20</td>
<td>62</td>
<td></td>
</tr>
<tr>
<td>Low right atrium</td>
<td>v 15</td>
<td>67</td>
<td></td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>230/20</td>
<td>58</td>
<td></td>
</tr>
<tr>
<td>Right ventricle</td>
<td>230/20</td>
<td>95</td>
<td></td>
</tr>
<tr>
<td>Pulmonary vein</td>
<td>a 25</td>
<td>13</td>
<td></td>
</tr>
<tr>
<td>Left atrium</td>
<td>v 15</td>
<td>13</td>
<td></td>
</tr>
<tr>
<td>Left ventricle</td>
<td>75/8</td>
<td>90</td>
<td></td>
</tr>
<tr>
<td>Ascending aorta</td>
<td>70/50</td>
<td>60</td>
<td>90</td>
</tr>
<tr>
<td>Descending aorta</td>
<td>80/50</td>
<td>70</td>
<td>88</td>
</tr>
</tbody>
</table>

Qp = 17.5 l/min per m²
Qs = 7.3 l/min per m²
PVR = 2.1 units m²
SVR = 7.6 units m².

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† F J Macartney is supported by the Vandervell Trust and the British Heart Foundation.
The mean frontal QRS axis was indeterminable with a clockwise loop.

The blood haemoglobin concentration was 13.4 g/dl. Cardiac catheterisation and angiocardiography was carried out. The haemodynamic data are recorded in the Table and demonstrate pulmonary hypertension, pressure in the right ventricle above the systemic, a right-to-left shunt at atrial level, and mild systemic arterial desaturation.

Angiocardiography was performed in both

Fig. 1 Left ventricular angiocardiogram in (A) frontal and (B) lateral projections, demonstrating connection of the left atrium to the smooth left ventricle (LV) by the mitral valve (MV). The pulmonary artery (PA) originates from the left ventricle.

Fig. 2 Right ventricular angiocardiogram in (A) frontal projection, early in the injection and (B) lateral projection, later in the injection. The catheter has been advanced from the right atrium through the tricuspid valve. The right ventricle (RV) is heavily trabeculated and hypoplastic. The aortic valve is atretic. However, the circumflex coronary artery (Co) fills from myocardial sinusoids, so that contrast medium passes from the coronary orifice (white arrow) to the ascending aorta (Ao).
Aortic atresia with transposition of the great arteries

ventricles and the ascending aorta through a retrogradely placed catheter. The films showed atrioventricular concordance (Fig. 1 and 2). There was aortic atresia, with the aorta connected to a hypoplastic right ventricle, and the pulmonary artery arising from the left ventricle. A hypoplastic ascending aorta was shown to fill through a large persistent ductus arteriosus. In addition, some opacification of the ascending aorta occurred from the right ventricle, not via the aortic valve, but via myocardial sinusoids communicating with the coronary arteries. The remainder of the systemic circulation was supplied by the descending aorta from the persistent ductus arteriosus.

A variety of surgical approaches was considered, including banding of the pulmonary arteries distal to the persistent ductus arteriosus, but eventually it was felt, in the light of her relatively symptom-free existence, together with the lack of a foreseeable method of complete repair, that surgical palliation was not indicated. Since then she has been followed-up in the outpatients' department and now, at the age of 6 years, remains reasonably well, attending special school. She continues below the third centile for height and weight. Her cardiac failure is moderately well controlled on digoxin therapy alone.

Discussion

The occurrence of aortic atresia in association with complete transposition of the great arteries has not previously been recorded.1 Our patient is extraordinary also in her continued survival and in the relatively good quality of her life. Survival to 6 years is far in excess of the average age at death which is 13.06 days in the common form of aortic atresia associated with ventriculoarterial concordance.1 The longest survivor with this more common connection is 3.5 years,2 though we have recently seen an 11-year-old boy with this condition. Though we cannot exclude the possibility that her survival is simply the result of an unusually large persistent ductus arteriosus and lack of restriction at the atrial septum,3 one cannot ignore the ventriculoarterial connection. It does appear that the left ventricle in this patient supports the systemic and pulmonary circulation far more effectively than does the right ventricle in typical aortic atresia with ventriculoarterial concordance.

References


Requests for reprints to Professor F J Macartney, The Hospital for Sick Children, Great Ormond Street, London WC1N 3JH.
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K M McGarry, J F Taylor and F J Macartney

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